ABSTRACT

TITLE: HEMANGIOPERICYTOMA AND SOLITARY FIBROUS TUMOR OF THE CENTRAL NERVOUS SYSTEM- A CLINICO-PATHOLOGICAL STUDY.

DEPARTMENT: Department of General Pathology**.**

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ABSTRACT

Background: Hemangiopericytomas (HPCs) and solitary fibrous tumours (SFTs) of the Central Nervous System (CNS) were classified as two distinct entities under meningeal based tumors in the 2007 WHO classification of tumors of the Central Nervous System. As both HPCs and SFTs of soft tissue have been found to share certain genetic alterations, these two tumors are now considered part of a single spectrum in soft tissue and as of 2016, also in the CNS. STAT6 and more recently GRIA2 have been reported as useful markers to identify these neoplasms.

The purpose of the present study was to carry out a detailed histological and immunohistochemical study of HPCs and SFTs of the CNS and to study the expression profile of STAT6 and GRIA2 in these tumors. The study also aimed to test the diagnostic capability of these two markers to differentiate HPCs/SFTs from their close histological and topographic mimics, meningiomas.

Methods: A detailed histological study of 75 HPCs and SFTs, reported between 1990 and 2014 was carried out. Immunohistochemistry was performed on these cases using rabbit monoclonal antibodies to STAT6 and GRIA2 A tissue microarray of 176 cases of meningiomas that served as a control, was also subjected to immunohistochemistry for STAT6 and GRIA2.

Results: There were 65 (86.66%) HPCs and 10 (13.33%) SFTs in the study population, which included 32 HPC WHO Grade II and 33 HPC WHO Grade III. The mean age at diagnosis of HPCs was 38 years and that of SFTs 45 years. Both HPCs and SFTs had a male preponderance with a M: F ratio of 1.32: 1 and 2.3: 1, respectively. The predominant architectural pattern in HPCs was of patternless sheets whilst in SFTs it was interlacing fascicles. Stromal collagen was seen in 90% of SFTs and in only 9% of the HPCs. Diffuse hypercellularity and nuclear pleomorphism were features almost exclusive to HPCs with hypercellularity seen only focally in 10% of SFTs. The mean mitotic count of SFTs was 1.5/10 HPF, whilst in HPCs, the mean mitotic count was 1.97/10 HPF for WHO Grade II HPCs and, 7.97/10 HPF for WHO Grade III HPCs. Nuclear immunopositivity for STAT6 was seen in 89.2% of HPCs, 80% of SFTs and none of the meningiomas. STAT6 immunohistochemistry had a sensitivity of 88% and a specificity

of 100% for the diagnosis of SFT/HPC. Cytoplasmic immunopositivity for GRIA2 was seen in 98.5% of HPCs, 100% of SFTs and 96.5 % of the meningiomas. GRIA2 immunohistochemistry had a sensitivity of 98.7% with a specificity of 3.5% for the diagnosis of SFT/HPC.

Conclusion: Since a high proportion of both HPCs and SFTs showed nuclear immunoreactivity for STAT6, the present study supports the merging of these two entities as one. The high specificity of STAT6 immunohistochemistry in diagnosing SFT/HPC makes it a useful marker to differentiate it from other dural based tumors such as meningiomas. On the other hand, GRIA2 with its low specificity is a poor marker for diagnosing SFT/HPC**.**